CWD Update 88

August 31, 2007

State and Provincial Updates

Illinois:

Paul Shelton, Illinois Department of Natural Resources provides the following: During July, IDNR identified a CWD-positive deer in LaSalle County after testing an animal showing classic signs of the illness. This was the first instance of the disease in this county. The deer was a 3 year old doe collected by a Conservation Police Officer after someone reported a sick, emaciated deer. The location was south of I-80, about 2 miles west of Grundy County, near the town of Seneca. This represents about a 25 mile distance from the previous southernmost positive in DeKalb County. Staff from the Division of Wildlife Resources are assessing the implications of the finding.

The total number of CWD-infected deer found in Illinois now numbers 189. Prior to this, the disease had been confined to Winnebago, Boone, McHenry, Ogle, and DeKalb counties. More than 28,000 deer have been tested in Illinois during the past 5 years. Illinois DNR CWD information is available at: http://dnr.state.il.us/cwd.

Editor's note: This finding in LaSalle County is a significant departure from the previously known distribution in Illinois. The new location is the first deer detected in the Illinois River basin, which winds southwest through Illinois towards St. Louis.

New Mexico:

Press Release from New Mexico Game and Fish (August 28, 2007):

LAS CRUCES: New Mexico recorded its 19th case of chronic wasting disease in deer in a sick animal found in the Bishop's Cap area of the Organ Mountains.

Officer Richard McDonald investigated a report of an emaciated deer July 12. The animal was unaware of human presence, chronically thirsty, urinating often, and staying in and near a water source. Officer McDonald followed the state's protocol for disease surveillance by killing the animal and sending it to the Veterinary Diagnostic Laboratory in Albuquerque for testing.

Based on the symptoms and the area from which the deer came, the laboratory was instructed that chronic wasting disease (CWD) was highly probable. Laboratory diagnostic testing confirmed presence of CWD in this deer. This is the 19th deer with confirmed CWD found since it was first detected in New Mexico in 2002. Two elk have also been found with CWD.

This deer was in Game Management Unit 19, where special CWD restrictions already exist for hunters.

Anyone who finds a deer or elk that appears unaware of human presence and displays symptoms including droopy ears, emaciation, chronic thirst, frequent urination, and reluctance to leave water, should report their observations to the Department of Game and Fish, Wildlife Management Division, (505) 476-8127.

New Mexico Game & Fish CWD information is at:
http://www.wildlife.state.nm.us/conservation/disease/cwd/index.htm.

Press Release is at:
http://www.wildlife.state.nm.us/publications/press_releases/documents/2007/082807releases.htm#CWD.

Recent Publications

Efficient In Vitro Amplification of Chronic Wasting Disease PrP^{RES}
Timothy D. Kurt, Matthew R. Perrott, Carol J. Wilusz, Jeffrey Wilusz, Surachai Supattapone, Glenn C. Telling, Mark D. Zabel, and Edward A. Hoover
Journal of Virology, September 2007, p. 9605-9608, Vol. 81, No. 17

Abstract: Chronic wasting disease (CWD) of cervids is associated with conversion of the normal cervid prion protein, PrP^{C}, to a protease-resistant conformer, PrP^{CWD}. Here we report the use of both nondenaturing amplification and protein-misfolding cyclic amplification (PMCA) to amplify PrP^{CWD} in vitro. Normal brains from deer, transgenic mice expressing cervid PrP^{C} [Tg(cerPrP)1536 mice], and ferrets supported amplification. PMCA using normal Tg(cerPrP)1536 brains as the PrP^{C} substrate produced >6.5 x 10^{9}-fold amplification after six rounds. Highly efficient in vitro amplification of PrP^{CWD} is a significant step toward detection of PrP^{CWD} in the body fluids or excreta of CWD-susceptible species.
http://jvi.asm.org/cgi/content/abstract/81/17/9605?etoc.

Ultrasensitive detection of scrapie prion protein using seeded conversion of recombinant prion protein
Ryuichiro Atarashi, Roger A Moore, Valerie L Sim, Andrew G Hughson, David W Dorward, Henry A Onwubiko, Suzette A Priola & Byron Caughey
Nature Methods - 4, 645 - 650 (2007)

Abstract: The scrapie prion protein isoform, PrP^{Sc}, is a prion-associated marker that seeds the conformational conversion and polymerization of normal protease-sensitive prion protein (PrP-sen). This seeding activity allows ultrasensitive detection of PrP^{Sc} using cyclical sonicated amplification (PMCA) reactions and brain homogenate as a source of PrP-sen. Here we describe a much faster seeded polymerization method (rPrP-PMCA) which detects greater than or equal to50 ag of hamster PrP^{Sc} (approximately0.003 lethal dose) within 2–3 d. This technique uses recombinant hamster PrP-sen, which, unlike brain-derived PrP-sen, can be easily concentrated, mutated and synthetically tagged. We generated protease-resistant recombinant PrP fibrils that differed from spontaneously initiated fibrils in their proteolytic susceptibility and by their infrared spectra. This assay could discriminate between scrapie-infected and uninfected hamsters using 2-mul aliquots of cerebral spinal fluid. This method should facilitate the development of rapid, ultrasensitive prion assays and diagnostic tests, in addition to aiding fundamental studies of structure and mechanism of PrP^{Sc} formation.
http://www.nature.com/nmeth/journal/v4/n8/abs/nmeth1066.html.

The following two articles are from the June 2007 edition (Volume 1772, Issue 6) of the journal Biochimica et Biophysica Acta (BBA) - Molecular Basis of Disease. This special edition of the journal, edited by Glenn Telling, is devoted to prion-related disorders and is available at: http://www.sciencedirect.com/science/journal/09254439.

Chronic Wasting Disease (review)
Christina J. Sigurdson and Adriano Aguzzi
Biochimica et Biophysica Acta 1772 (2007) 610–618

Abstract: Until recently, chronic wasting disease of cervids, the only prion disease affecting wildlife, was believed to be geographically concentrated to Colorado and Wyoming within the United States. However, increased surveillance has unveiled several additional pockets of CWD-infected deer and elk in 12 additional states and 2 Canadian provinces. Deer and elk with CWD have extensive aggregates of PrP^{Sc} not only in the central nervous system, but also in peripheral lymphoid tissues, skeletal muscle, and other organs, perhaps influencing prion shedding. Indeed, CWD is transmitted efficiently among animals by horizontal routes, although the mechanism of spread is unknown. Genetic polymorphisms in the Prnp gene may affect CWD susceptibility, particularly at codon 225 (S/F) in deer and codon 132 (M/L) in elk. Since CWD infects free-ranging animals and is efficiently spread, disease management will be a challenge.

Motor behavioral and neuropathological deficits in mice deficient for normal prion protein expression
Karah E. Nazora, Tanya Sewarda and Glenn C. Telling
Biochimica et Biophysica Acta 1772 (2007) 645–653

Abstract: It has been difficult to reconcile the absence of pathology and apparently normal behavior of mice lacking prion protein (PrP), referred to as $Prnp^{0/0}$ mice, with a mechanism of prion pathogenesis involving progressive loss of PrP^{C}-mediated neuroprotection. However, here we report that $Prnp^{0/0}$ mice exhibit significant age-related defects in motor coordination and balance compared with mice expressing wild type Prnp on a syngeneic background, and that the brains of behaviorally-impaired $Prnp^{0/0}$ mice display the cardinal neuropathological hallmarks of spongiform pathology and reactive astrocytic gliosis that normally accompany prion disease. Consistent with the appearance of cerebellar ataxia as an early symptom in patients with Gerstmann–Sträussler–Scheinker syndrome (GSS), an inherited form of human prion disease, motor coordination and balance defects manifested in a transgenic (Tg) mouse model of GSS considerably earlier than the onset of end-stage neurodegenerative disease. Our results are consistent with a mechanism in which loss of normal PrP^{C} function is an important pathological component of prion diseases.